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Floating Thrombus in the Ascending Aorta of the Patient with Systemic Sclerosis

— A case report —

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Aortic thrombi are important because it can cause the central and peripheral embolizations. Aortic thrombi can occur anywhere in the aorta but extremely rare in ascending aorta without atherosclerosis, aneurysm, cardiosurgical or traumatic state. Systemic sclerosis (SSc) is an autoimmune disorder of connective tissue and it can involve multisystem. Enhanced coagulation pathways, decreased fibrinolysis, and endothelial dysfunction probably contribute to vascular events in SSc. We report a case of a highly mobile thrombus in the ascending aorta, presented as an acute embolic stroke in the patient with systemic sclerosis. Surgical removal was performed to prevent recurrent embolic events.

Key words: 1. Aorta
2. Thrombosis
3. Systemic sclerosis

CASE REPORT

A 67 year old woman was hospitalized in the emergency room due to dysarthria started 4 hours before hospitalization. The patient was diagnosed with systemic sclerosis in 2003 and was taking methylprednisolone 2 mg per day at our clinic's rheumatic internal medicine department. According to physical examination findings, sensorium was normal where as dysarthria, facial palsy, and right deviant of tongue were observed. Blood pressure was 140/80 mmHg, pulse was 102 beats per minute, respiratory rate was 20 times per minute, body temperature was 36.9°C, and there were no abnormalities in chest auscultation. According to blood test result, number of white blood cells was increased and platelet level was normal. Cardiomegaly was observed through simple chest X-ray and acute infarction was observed around the large sulcus and parietal lobe through MRI (Fig. 1). From 3-D CT scan of carotid artery and chest to find out the cause of cerebral infarction, lesion suspicious of aortic arch thrombus was observed. In enhanced image, it was 1.8 cm-long radiolucent (Fig. 2). In transthoracic echocardiogram, normal cardiac function was observed, no abnormal observations were found in regional wall motion and heart valves, and floating thrombus was found in ascending aorta (Fig. 2). According to hematological test, anti-cardiolibin Ab-IgG level was 2.0 GPL U/mL (normal: less than 9.9), anti-cardiolibin Ab-IgM level was 1.0 MPL U/mL (normal: less than 6.9), lupis anticoagulant (−), protein S antigen was 114% (60～150), protein S activity was 93% (58.7～119.2), antiphospholipid Ab IgG was 1.0 U/mL (less than 9.9), and antiphospholipid Ab

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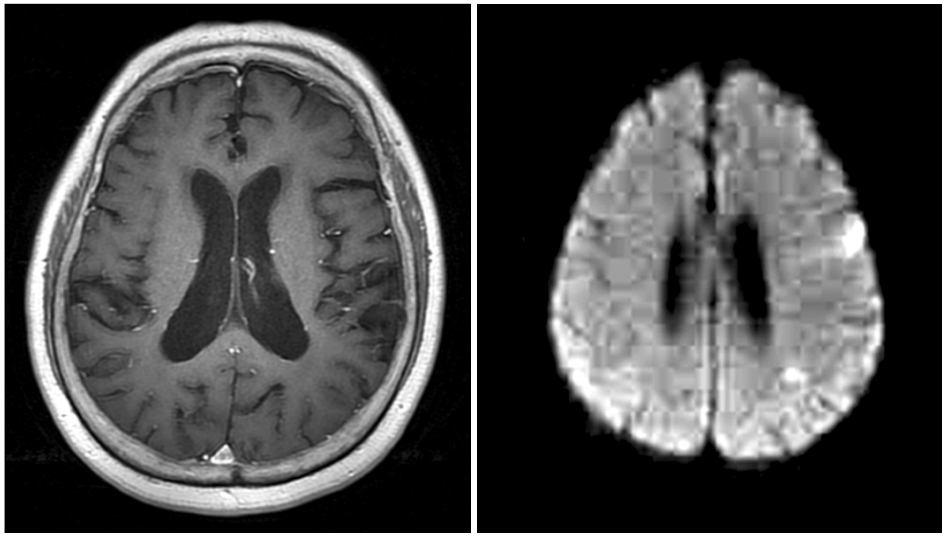

Fig. 1. Brain MRI shows acute infarction in left precentral gyrus and left parietal lobe.

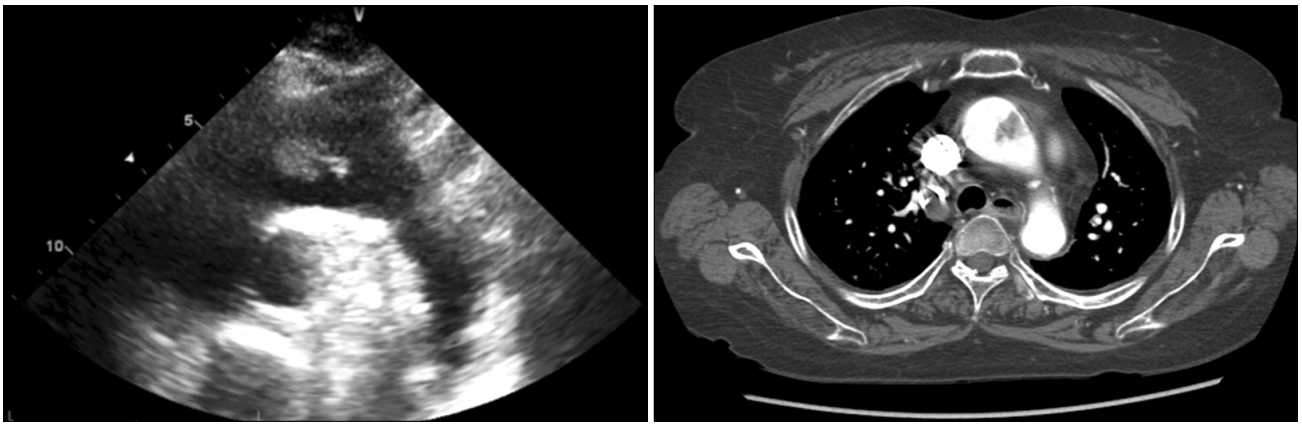

Fig. 2. Preoperative TEE and 3D thoracic aorta CT show thrombus in the ascending aorta.

IgM was 2.0 U/mL (less than 9.9) showing normal values. However, protein C activity was 167% (70～130) and protein C antigen was ＞160% (72～160) showing increased levels. Due to the facts that size of the thrombus was large, location was from ascending aorta to aortic arch, thrombus was highly mobile and cerebral embolism occurred, surgical treatment was executed rather than a medical approach in removing the thrombus. Median sternotomy was done, arterial cannula was inserted through 8 mm vascular graft anastomosed in right axillary artery, venous cannula was inserted in superior and inferior vena cava, started extracorporeal circulation, and decreased central temperature to 22°C. Cardiac arrest was induced by inserting retrograde cardioplegic solution in coronary sinus, right innominate artery and left common carotid artery were cross clamped, and selective antegrade cerebral perfusion through right axillary artery was done eventually leading to circulatory arrest. Incision was made in ascending aorta and 2.5 cm-long intraaortic thrombus was completely removed (Fig. 3). There were no abnormalities in intima of ascending aorta where the thrombus was attached. Aortic incision was sutured, right innominate artery and left common carotid artery were unclamped resuming systemic circulation, and weaning from cardiopulmonary bypass was done without complications. Selective antegrade cerebral perfusion was done for 15 minutes and cardiopulmonary bypass was done for 128 minutes. Based on pathological test of the thrombus, there were no evidence of malignant neoplasm, and the patient recovered without complications and was discharged.

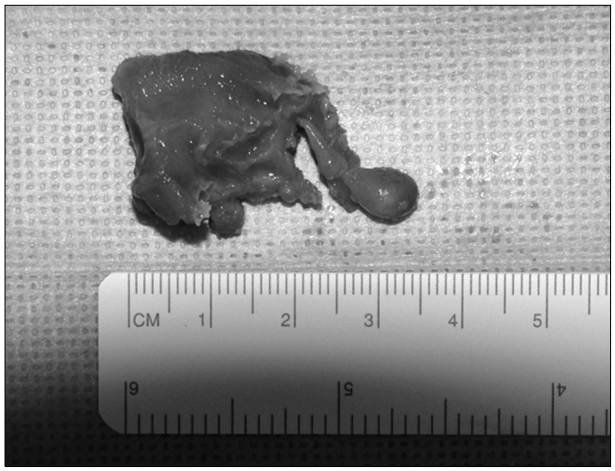

Fig. 3. Gross photo of about 2.5 cm-sized thrombus.

DISCUSSION

Factors related with arterial thrombus are arteriosclerosis, arteriodissection, trauma, malignant tumor, and hemostatic disorder [1,2]. In this case study, the patient had systemic sclerosis from several years ago and was taking methylprednisolone 2 mg per day. There are no clearly known causes of systemic sclerosis, but natural death of cell caused by anti-endothelial cell antibody is suggested as a hypothesis. This usually affects microvessel causing proliferation of endothelial cell, medial thinning, and thickening of basilar membrane. Eventual destruction of vessel causes angiodyskinesia, functional disorder of endothelial cell, thickening of vessel wall, activation of coagulation factor, reduction of fiber disassociation, and increase in attachment of platelets and lymphocytes [3]. In cases with systemic sclerosis, thrombus in extremity is relatively common and floating thrombus inside the aorta is known to be rare [1].

Thrombus mostly occurs in elder patients, is usually stationary forming atherosclerotic plaque, and often occurs in extremity [1,2]. Patient of this case did not have past history that is known to cause thrombus such as artery detachment, trauma, or malignant tumor. Evidences of infection malignant tumor, arteriosclerosis, and inflammatory change were not found after postoperative pathological test of specimen. The patient did not smoke, had no hypertension or diabetes, did not show high parameters in blood test, but only showed higher levels of protein C activity and protein C antigen than normal. It is difficult to prove direct relationship between systemic sclerosis and aortic thrombus. However, long-term use of steroid and vessel damage due to systemic sclerosis might be related to thrombus formation [1]. Thrombus occurring in patients with systemic sclerosis is relatively common in extremity. Since occurrence in ascending aorta is rare, this case can be considered as meaningful.

Floating thrombus inside the aorta can cause systemic embolism. Thus, fast and exact diagnosis and treatment is necessary. Transesophageal echocardiography is non-invasive and easy method used widely for diagnosing thrombus in heart or aorta. Additional information can be achieved through CT angiography [1,2,4].

There are several different methods in aortic thrombus treatment such as anticoagulant therapy, thrombolytic therapy, aspiration thrombectomy, and thrombectomy, but there are no evidence that which method is better [1,2,5]. In medical treatment, thrombolytic agent and anticoagulant are used. After using anticoagulant with heparin for several days, size reduction of thrombus must be observed and heparin treatment must be maintained until the thrombus is completely gone. In general, when the size of thrombus is large and thrombus is mobile, surgical removal is preferred as primary treatment because of the risk factors such as systemic embolism, repeated embolism, and medical treatment failure [1,2,5]. In this case, heparin was used for 2 days after cerebral infarction prior to surgery. Thrombectomy was executed since it was highly mobile, large in size, did not change in its size through follow-up ultrasonic test, and repetitive cerebral embolism was possible. This report is written in order to inform a rare case where thrombus in ascending aorta of a patient with systemic sclerosis was surgically treated.

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